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A RARE PRESENTATION OF APLAS

Rheumatology

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ABSTRACT

- APLAS/ APS is an autoantibody mediated acquired thrombophilia with recurrent arterial le venous thrombosis and pregnancy morbidity.
- It primarily affects females . 5 cases per 1 lakh population .
- Diagnosis of APLAS should be considered in cases of arterial or venous thrombosis, cardiovascular accidents in individuals less than 55 years of age.
- We reported a case of 17year old un married female with history of Abdominal pain associated with abdominal distension since 3 months diagnosed with BUDD CHIARI SYNDROME .
- She was screened and evaluated for Autoimmune diseases and found to have Antiphospholipid antibody syndrome.
- She was kept on Anticoagulants and beta-blockers and diuretics.
- Patient was doing well on follow up.
- Any arterial / venous thrombosis events in young female needs active investigation so that appropriate treatment can be started to halt the disease process.
- **BACKGROUND**
- APLAS/ APS is an autoantibody mediated acquired thrombophilia with recurrent arterial le venous thrombosis and pregnancy morbidity.
- It primarily affects females . 5 cases per 1 lakh population .
- Diagnosis of APLAS should be considered in cases of arterial or venous thrombosis, cardiovascular accidents in individuals less than 55 years of age.
- The diagnosis of APLAS include clinical and laboratory criteria
- Presence of at least one clinical and one laboratory criterion is compatible with diagnosis.
- Anti cardiolipin antibody, lupus anticoagulant and anti B2 GP1 antibodies can be positive among APLA positive individuals on tow occasions 12 weeks apart. (LABORATORY CRITERIA)
- Vascular thrombosis/ pregnancy morbidity (CLINICAL CRITERIA)

KEYWORDS

CASE REPORT

- A 17 year old female by name B. Balamma , resident of prakasam district presented to OPD with complaints of abdominal pain in right hypochondrium associated with abdominal distension since 3 months , insidious in onset and progressive. No yellowish discolouration of eyes. No pedal edema. No history of blood transfusions.
- On examination, she had hepatomegaly and moderate ascites with no signs of liver cell failure.

LABORATORY INVESTIGATIONS

- Complete blood count

Hb 11.6 mg/dl
TRBC 4.2
TWBC 8000
P 65% L 30% M02%
ESR 20
PC 2,80,000
PCV 42
Peripheral smear – Normal study

- Liver function tests , Renal function tests, s electrolytes- Normal study
- Serum beta GP1 antibody – 62units/ml
- Anticardiolipin antibody IGM- 26 (positive)
- Anticardiolipin IGG – 64(positive)
- Lupus anticoagulant screen -68 seconds , Control- 31 seconds
- Lupus screen / control – 2
- Thyroid profile – T3 1.27 ng/ml
  T4 10.24ug/dl
  TSH 3.5 uIU/ml
- Serum ANA levels – 1.28
- ANA profile – Negative
- USG abdomen Doppler shows –

Enlarged liver (17 cms) with normal echo texture
Absent blood flow in right and middle hepatic veins with narrowing of ostia near IVC
Moderate ascites
Features suggestive of BUDD CHIARI SYNDROME.

- UGI endoscopy – Grade 2 esophageal varices with mild portal gastropathy.
- Impression – PORTAL HYPERTENSION

DISCUSSION

- Patient was advised followup to OPD regularly. Entire investigation profile was repeated again.
- Patient was diagnosed to have Budd chiari syndrome secondary to APLAS
- Catastrophic APLAS is a life threatening rapidly progressive thromboembolic disease involving simultaneously 3 or more organs .
- One third of patients with SLE or other autoimmune diseases possess these antibodies with only 5-10% developing APLAS.
- Clinical manifestations represents the consequences of arterial or venous thrombosis.
- Treatment options include aspirin 80mg , Low molecular weight heparin, oral anticoagulants like warfarin, Rivaroxaban, fondaparinux and IV immunoglobulins.

CONCLUSION

Young females with arterial or venous thrombosis needs to be evaluated for congenital or acquired thrombophilias(APLAS) to prevent further thrombotic events .

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CONFLICTS OF INTEREST- NIL
CONSENT – consent was taken

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